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The Annual Meeting of the International Skeletal Society (6th)

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THE SIXTH ANNUAL MEETING OF THE INTERNATIONAL SKELETAL SOCIETY

The Sixth Annual Meeting of the International Skeletal Society was held in Bavaria's late summer sunshine and amidst Munich's typical Gemütlichkeit. The Society is a new and dynamic one, organized by radiologists specializing in bone diseases, but including in the membership prominent pathologists and orthopedic surgeons. The diagnosis of bone disease is somewhat unique in comparison with other subspecialities of radiology as it occupies both ends of a difficulty spectrum and not much middle ground. The great majority of bone radiographs are simple, straightforward examples of fractures and dislocations. However, at the other end of the spectrum are multiple types of dwarfism, inborn errors of metabolism, derangements of calcium metabolism, as well as obscure benign and malignant bone tumors. It is, of course, the latter end of the spectrum that concerns the International Skeletal Society.

The six-day meeting was divided into two halves: the first, a session closed to all but members of the International Skeletal Society (this writer the only exception) and the second, a symposium and refresher course open to all. In the first half of the meeting the members of the society met to discuss common problems, interesting cases and new developments. During the second and open portion of the meeting, the members of the society acted as faculty, presented papers and held symposia for the audience.

The closed meeting consisted of papers alternating with groups of case presentations. This format was a good one for maintaining audience interest. The first paper was by William F. Enneking. (Dept. of Orthopaedic Surgery, University of Florida, Gainesville). Enneking's paper was one of two on the "Newer Approaches to the Surgical Management of Malignant Bone Tumors." He spoke specifically about resection and reconstruction for primary malignancies of the pelvic innominate bone. Generally speaking, pelvic surgery is hazardous, difficult to carry out, and deforming in the extreme for the patient. Some malignant bone tumors of the pelvis are best treated by hemipelvectomy, some by radiation therapy but others lend themselves to resection of a portion of the bone with reconstruction. Enneking's criteria for resection and reconstruction are: 1) the tumor must be radioresistant; 2) the patient must be free of distant metastatic disease; 3) it must be anatomically feasible, and 4) the patient must have the right personality to undertake the necessary rehabilitation. On the other hand hemipelvectomy is carried out if resection and reconstruction will result in: 1) inadequate function; 2) excessive shortening of the leg; 3) instability; 4) prolonged povalescence; or 5) inadequate tumor margins. Reconstruction surgery attempts to preserve functions of the leg and falls into three types: 1) high resection of the innominate bone, 2) resection of the acetabular portion of the innominate bone, and

3) resection of the pubic and ischial portions of the bone. Enneking operated on 57 patients, 25 of whom required a hemipelvectomy but for the remaining 32 resection and reconstruction were carried out. Of these 23 had a good result, 4 satisfactory, and 5 poor. The results speak for themselves and are particularly praiseworthy when one considers the disease.

A related paper by Bertil Stener, (Dept. of Orthopaedic Surgery, Goteberg, Sweden) was concerned with high amputation of the sacrum for malignant bone tumors. Stener described two operations, one between the first and second sacral segments that preserves the S-1 nerves and the second through the body of S-1, sacrificing the first sacral nerves. The author demonstrated his surgery with photographs as well as radiographs and line drawings which made this technical surgery clear even to a neophyte. The author also carried out experimental pelvic stress measurements on cadavers. The pelvis was submitted to a downward force from above the 5th lumbar vertebra. found that in comparison to unresected specimens, if the resection was through the first sacral segment, there was a 50% weakening, and if the resection was between the first and second sacral segments only a 30% weakening. Stener concluded from this work that after resection considerable residual strength remained in the pelvic ring and that it was adequate for full weight bearing and early ambulation of these patients in the post-operative state.

In a very interesting paper from South Africa, P. Beighton (Dept. of Human Genetics, Capetown) described an inherited, progressive and lethal disorder of bone he has named sclerosteosis. The author studied 45 patients, all of whom are Afrikaaners. The disease consists of progressive thickening and enlargement of many bones but particularly the skull with compression of the brain, gigantism and early facial palsy. The bones become very dense and resistant to trauma. (Beighton described one case in which a factory worker with sclerosteosis was hit in the jaw by a fly wheel with sufficient force to fragment the wheel but his jaw remained intact.) In addition 90% have syndactaly. The disease is autosomal-recessive and almost entirely confined to South Africa. It is of considerable interest that van Buchem's disease is quite similar, but not as severe, and all 15 of Van Buchem's patients are Dutch, while Beighton's patients are Afrikaaners all of whom are of Dutch descent. Beighton has been searching for radiographic clues in the carriers of the recessive trait but thus far has not been successful.

W. Remagen (University of Basel, Switzerland) discussed "Electron microscopic findings in fibrous dysplasia." The author demonstrated with electron photomicrographs abnormal microfibrils of collagen that give rise to the spotty calcification charasteristic of fibrous dysplasia. Normal collagen fibrils, which may be immediately adjacent to the microfibrils, produce normal bone. The author believes that the microfibrils are due to an enzyme deficiency which inhibits the

formation of normal collagen.

Ronald Murray (Royal National Orthopedic Hospital, London) read a paper on the "Instability of the symphysis pubis and its relationship to osteitis condensans ilii." The latter is an increase in bone density in the interior-anterior angle of the ilium adjacent to the sacrum. Although this benign condition of women in the reproductive age group has been recognised for years, its exact etiology has been unknown. Recently, a relationship to hormonal ligamentous relaxation during pregnancy has been proposed along with a better name, stress sclerosis. Murray related stress sclerosis to instability of the symphysis pubis known to be caused by ligamentous relaxation during pregnancy. Relaxation and instability of the symphysis pubis after pregnancy is always seen concurrently with stress sclerosis and both are more common in women who also do manual labor during pregnancy. Murray also described instability of the symphysis pubis secondary to athletic stress. He radiographically examines the pelvis with the patient first standing on one leg and then the other in order to demonstrate the degree of instability at the symphysis.

Resorption of corticle bone is seen in a number of different diseases, the most common of which is chronic renal failure. E. Meema (Department of Radiology, Toronto Western Hospital, Toronto, Canada) described work he has done on microradioscopic quantitation of endosteal and periosteal bone resorption. Using hand radiographs of 60 normal individuals and 60 patients with chronic renal failure, made with Kodak M industrial film for very fine detail and ordinary occular magnification, Meema established an index between the number of striations in a proximal phalynx and in the metacarpal of the same digit. A large majority of patients with the chronic renal failure fall outside the normal index. In addition, by using the same measurements he showed that therapy for these patients with 1,25 (OH) 2 vitamin D3 was not helpful in preventing endosteal erosion but did improve intracortical and periosteal resorption.

I.P.C. Murray (Randwick, Australia) discussed "The Effect of Hypercalcemia on skeletal scintigraphy." About 60% of patients who have died with various hypercalcemic conditions will demonstrate histological viseral calcification predominently lungs and stomach, but only about 3% can be seen by x-ray examination or by "mTc bone scans. The author believes that the hydroxy-apatite deposited in the organs affected is labeled with the isotope. However, the condition is thought to be temporary and in a case he described lasted less than 72 hours. In reviewing the literature the author found 12 patients with chronic renal failure, primary hyperparathyroidism or hypervitamosis D and 15 patients with various malignancies who demonstrated visceral calcification by 99mTc scanning.

It may be quite difficult even for an expert bone pathologist to distingish cell type and at times even between benign and

malignant bone tumors. Frequently a panel of experts will disagree among themselves. Aggressive non-metastasing benign bone changes can be confused with malignant growths and radical surgery carried out erroneously unless great care is taken. To aid in this regard C.P. Adler (Freiburg, West Germany) described a pilot study involving the cytophotometric measurements of various bone tumors. This new method allows the pathologist to measure the DNA content of cell smears taken from the tumor. The DNA content increases considerably from the diploid to the tetraploid phase of mitosis. The author constructed histograms and was able to distinguish between the great majority of benign and malignant bone tumors. A few benign neoplasms, which tend to be recurrent after excision, fall slightly above the remainder of the benign tumors but still can be classified as benign.

M. Becker (New York Univ., New York) described the latest statistics regarding the pathological changes in newborn infants resulting from warfarin (Coumadin) therapy in the mothers. Warfarin is a vitamin K antagonist sometimes prescribed during pregnancy for thrombosis. If taken during the first trimester of pregnancy, approximately 8% of the newborns develop the syndrome called Chondrodysplasia punctata, a much smaller percentage than originally thought. However, warfarin therapy at any time during pregnancy may increase bleeding tendancy in the newborn.

Another sophisticated use for the the CT Scanner was described by H. Genant (Univ. of California, San Francisco, CA). He outlined a method of measuring bone mineral content by the CT attenuation coefficients. Corrections have to be applied for scanner drift with standard solutions and care must be taken to measure the vertabrae at precise points. In addition, CT attenuation coefficient measurements of bone marrow were made comparing normal cases with abnormal ones in which the marrow was packed with metastatic neoplasm.

The second portion of the meeting, the refresher course, dealt largely with basic concepts and new classifications based on recently gained knowledge. The first of these papers by Hans-Georg Willert (Univ. of Göttingen, Göttingen, Germany) was concerned with the morphology and classification of congenital bone defects. His work was stimulated by the Thalidamide catastrophe during which so many children, whose mothers were on the drug, developed deformities. Evidently the action of Thalidamide was at a very early stage of embryogenesis and affected sensory nerve and sclerotome development, which, in turn, adversely affected the normal maturation of the limbs. Willert has developed a transverse and longitudinal classification based on the study of a number of Thalidamide babies. He demonstrated his material with radiographs, line drawings, and photomicrographs of the gross specimens. All the patients had several features in common. 1) Hypoplasia and incomplete separation of the various hypoplastic portions, 2) delay in ossification,

3) loss of normal limb shape.

Jürgen Spranger (Mainz, Germany) has reclassified the congenital abnormality, Osteogenesis Imperfecta, based on the genetic pattern; autosomal-dominant or autosomal-recessive, on the severity of the bone disease, and on the presence or absence of a blue sclerae.

Herbert J. Kaufmann (Paris, France) showed, with multiple radiographs as well as ultrasound examinations, how the diagnosis of various skeletal dysplasias can be made while the fetus is still in the uterus. He emphasized that the intrauterine diagnosis requires radiographs made in the proper projection and an extensive knowledge of the various types of congenital bone disorders that one might encounter.

A paper by H.K. Genant (Univ. of California, San Francisco, CA) was concerned with the use of scintigraphy (bone scanning) for non-neoplastic bone and joint disorders. The isotope 99mTc is absorbed or exchanged for another ion on the hydroxy-apatite crystal. This absorption or exchange is influenced by 1) blood flow, the state of osteogenesis, 3) the surface area, 4) capillary permeability, and 5) extra-cellular fluid (ecf) concentration. Patients with sacroileitis as part of their joint disease were examined and the radioactive isotope uptake measured over the sacroiliac joints was compared to that of the sacrum and graphs constructed. In normal patients, the uptake by the sacrum is a approximately the same as by each sacroiliac joint. In comparing the normal controls with the patients he found that the test was not significant for all grades of sacroileitis but only for early, active disease. In other words, if grades 3 and 4 sacroileitis which represent an older, more advanced but often less active process were eliminated, the test was quite significant in separating normal patients from those with early disease.

In the second part of his paper, Genant described how scintigraphy can also be used for the detection of loosened prostheses or infection in patients with a total-hip replacement. Post-operative pain is not an uncommon complication of a joint prosthesis. The problem is to separate those with loosening of the prostheses or infection from patients with other causes of pain. Scintigraphy has proved very useful in this regard, moreover, radioactive uptake at the distal tip of the prosthesis was particularly sensitive and specific for loosening or infection.

The use of arthrography for diseases of the hip was thoroughly reviewed by Amy Beth Goldman (Hospital for Special Surgery, New York). Goldman initially discussed hip diseases and arthrography in infant and children, and secondly in adults. In children she showed cases of 1) infection, 2) congenital hip dislocation vs. subluxation, 3) detection of an unsuccessful reduction of a hip

dislocation, 4) the diagnosis of Legg-Perthes disease, important not only in itself but to determine whether surgery will be necessary. In adults, Goldman first described various arthritides and how not only the type but the extent of arthritis can often be seen by hip arthrography. Other pathology shown in the adult were trauma and the evaluation of hip prostheses. Two percent of all prostheses become infected and 100% become loose in 10 years. Therefore, hip arthrography is also important in this very common surgical procedure.

The second day of the refresher course opened with a discussion of various bone tumors by four emminent American pathologists. The papers given and the discussion which followed was largely a visual description of the histology and radiologic appearance of a number of different tumors. The point was well made, however, throughout the bone tumor papers, that the histology alone often does not provide the final answer and that a final diagnosis can only be arrived at by a correlation of the radiologic appearance, the clinical course of the disease, the biopsy site as well as the histology. Bone pathology is unique in this regard and many tumors have been classified differently after good correlation and a second biopsy. Therefore, not only is the radiologic appearance of the neoplasm itself critical but the radiologist must direct the surgeon as to the best biopsy site. The surgeon for his part must be sure that adequate tissue is provided for pathological examination. A good bone pathologist only then makes a diagnosis in conjunction with the radiologist's impression and the surgeon's findings.

Rodney Bluestone (Wadsworth Veterans Administration Hospital, Univ. of California) delivered a paper entitled "The Seronegative Spondyloarthropathies." The seronegative arthropathies are those in which serological tests for circulating abnormal antibodies are negative. Seropositive arthropathies, rheumatoid arthritis, for example, attack the large synovial joints, while the seronegative arthropathies such as ankylosing spondylitis tend to involve the cartilaginous joints first and later the smaller synovial joints of the spine. These inherited disorders result in chronic inflammation and instead of destruction and sub-luxation lead to post inflammatory fibrosis and eventual joint fusion. In the spine the fusion is often periarticular as the inflammed soft tissues around the joints later calcify and fuse. In addition to ankylosing spondylitis, with or without colitis, Reiter's syndrome, postsalmonella, post-yersinia and psoriatic arthritis fall into this category.

An excellent paper describing the anatomic basis of a number of different radiographic observations of the proximal femur was delivered by M. Pitt (Univ. of Arizona, Tucson, Arizona). The title "Lines and Lucencies of the Proximal Femur" did not do justice

to the depth of the work carried out. Pitt used Indian skeletal remains found in the Arizona desert, radiographs, as well as anatomic specimens to analyze in detail normal radiographic observations of the proximal femur. An anatomic basis for these well-known radiographic observations had not been described previously and this work, meticulously carried out, will be of considerable help to the radiologist who must distinguish between early pathology and normal anatomy.

M. Kricun (Hahnemann Hospital, Philadelphia, PA) elaborated on the "Significance of Red bone marrow distribution" throughout the body at various ages. Red bone marrow is blood cell producing marrow as opposed to yellow or fatty marrow. In the embryo, the blood cells are produced by other organs such as the liver and spleen and it is only after birth that all the blood elements are produced in the bone marrow. The newborn has red bone marrow throughout the bones of the body, which progressively decreases, beginning with the small bones of the hands and feet, to age 25. Kricun then related the various neoplasms of bone marrow such as multiple myeloma, reticulum cell sarcoma, and Ewings sarcoma to the presence of red marrow in the affected bone at a certain age. Put another way, specific malignancies appear only in bones that contain red bone marrow, which in turn accounts for the age distribution commonly seen among the various malignancies. Kricun went on to describe the vascular anatomy of bone and related this to the deposition of metastases. In concluding his discussion Kricun noted that multiple myeloma, for example, appears almost exclusively in the axial skeleton and in the proximal portions of large bones all of which contain red marrow. If, for any reason, such as a chronic anemia, there is an increase in red bone marrow with a redistribution toward the periphery, then the distribution of lesions of myeloma will follow.

In one of the final papers K. Subbaro (Montefiore Hospital, New York) described the changes of skeletal fluorosis in certain endemic areas of the world. Fluorosis is the biochemical replacement of CaCO3 with CaF2. Industrial fluorosis also occurs but it is never as severe as the endemic variety. Excess flouride causes white patches and yellow lines on the enamel of the teeth and a generalized increase in bone density. Bone density and weight at autopsy have been measured up to 3 times normal. Although this is a pathological condition and can cause spinal osteostenosis, it has not been known to shorten the normal life span. In endemic areas diseases that increase the intake of water increase the severity of the fluorosis. The flouridation of city water supplies to decrease tooth decay is approximately one part per million and does not cause the disease. The pathological changes in the teeth and bones are not only related to an increased amount of flouride in the water, but to the time of exposure and the amount of water ingested.